on a case of chronic sinusitis, the virulency of the bacteria is greatly increased, owing to the wound secretions forming an excellent nutrient medium; hence the cellulitis or erysipelas which develops after some of our external operations in cases of chronic nasal accessory sinus disease. I am not surprised that a small percentage of patients having chronic sinusitis, on whom intranasal operations are performed, develop acute otitis media or even mastoiditis. The wonder is that more do not.

THE METABOLIC CHANGES IN HEMATOPORPHYRINURIA NOT OF DRUG ORIGIN.

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Hematoporphyrin is simply hematin without the iron. It is found in the normal urine in faint traces (Garrod). This amount is increased, though not enough to color the urine, in various morbid conditions, such as measles and rheumatic fever. It has no clinical importance so far as we know in these minute amounts. When it occurs in sufficient amount to color the urine, causing hematoporphyrinuria, the act is significant and often ominous.

Hematoporphyrinuria is best known in connection with the use of sulphonal, trional, and tetronal. But besides these drug cases, the condition occurs as the result of various other toxemias.

T. K. Monroe[1] reports a case of periodical vomiting in a boy, aged seven years, occurring about every two months, and each attack lasting about a fortnight, accompanied with hematoporphyrinuria and acetonuria. The attacks continued for two years, and the patient finally died of the disease. He reports, also, the case of a woman, aged thirty-six years, who suffered from multiple neuritis and a pelvic exudate, accompanied with hematoporphyrinuria. She recovered in about seven weeks. Also a third case, that of a woman, aged twenty-six years, who suffered from hematoporphyrinuria after having become chilled while bathing at sea. In these cases there was no drug poisoning and no internal hemorrhage.

W. Langdon Brown and H. O. Williams[2] report the case of a married woman, aged twenty-two years, who had three attacks

[1] Quarterly Journal of Medicine, 1907-08, i, 49.

[2] Lancet, 1909, i, 1105.

of abdominal pain and vomiting with hematoporphyrinuria, lasting several weeks. In the last attack she had three epileptiform fits and became delirious and drowsy, but finally recovered. The authors collected other cases of hematoporphyrinuria, not due to drugs, but associated with gastro-enteric disturbance and apparently due to intestinal toxemia.

From the reported cases it would seem that hematoporphyrinuria, when not due to drug poisoning, is due, therefore, to some toxemia or to some internal hemorrhage.

The following case does not surely belong to any of these groups It is reported because of the exceptional opportunities which were furnished me for making blood and metabolism studies in connection with the progress of the case, through the courtesy and helpfulness of Dr. T. C. Roper, of the New York Hospital Laboratory, and of Dr. T. W. Hastings and Dr. Warren, of the Cornell University Medical College Laboratory.

Summary. Male, aged sixty-seven years. Apoplectic seizure with temporary hemiplegia, short excited and long confusional mental state, lasting about three months. Development of marked hematoporphyrinuria, lasting for two months. Seven nitrogen examinations, showing increase of kreatinin and rest nitrogen; no signs of hemolysis, no drug intervention, no known intestinal toxemia. Hematoporphyrinuria is evidence of a metabolism disorder, and not a sign of a serious blood condition.

Mr. J. T. J., aged sixty-seven years, married, two children; occupation, president and financier. The patient has been a very strong, rugged, hard-working man, never taking a vacation. He has never drunk alcohol in any form, nor tea or coffee; he has not used tobacco nor any drugs. His food habits are said to have been particularly intemperate. He would eat all kinds of food, including meat, in very great amount. About twelve years ago he had some heart trouble, for which he was sent to Nauheim for two successive years. He was relieved by these trips. He had an attack of vertigo in January, 1909. He recovered from this and continued his work.

On February 23, 1909, while at work in his office, he became confused and was unable to talk connectedly. An hour or so later he could not talk at all, and on attempting to do so would burst out crying. On attempting to walk, he was found to be paralyzed on the right side. He was placed in bed and slept during the night. The next day he tried to get up and dress, but was unable to do so on account of his right hemiplegia. He tried to talk, but failed, and his efforts would end in explosive crying. He had also some difficulty in swallowing. He was just able to sign his name to a power of attorney.

He improved slightly in the next week, and then went into a semicomatose state, which lasted for two days, during which period he would at times arouse himself and become delirious. He came out of this coma and was then more quiet, but was confused and at times delusional, thinking that he was being persecuted and poisoned. He could speak now a little, but his speech was very indistinct, and he could not sit up or stand without help.

He again gradually improved and in about a month after the beginning of the attack he was able to be brought to New York. At that time he could talk a little and could sit and stand, but the fatigue of the journey set him back; so that when he reached here he was again in a condition of semicoma, from which he would occasionally arouse and become delirious—trying to get out of bed and showing evidences of hallucinations of sight and hearing.

I saw him first on March 25, two days after he reached the city. He had then quieted down somewhat, and responded to simple questions and recognized members of his family.

On examination, he showed absolutely no signs of a hemiplegia. His right arm and leg were as strong as the left. There was no change in the reflexes, either superficial or deep. There was no Babinski reflex; no ankle clonus; no disturbance of deep or superficial sensation. His only peculiarity was a tendency to hold the left hand in whatever position it was placed—a sort of catatonic state. His special senses were apparently normal, though his mental condition prevented careful examination. The temperature was normal; the heart sounds good; the pulse 70; the blood pressure nearly 200 mm. Hg, and but a little higher on the left than on the right. He had for three days Cheyne-Stokes respiration; the lungs were normal. The urine showed a trace of albumin and some casts; the amount was normal.

During the next few weeks his condition slowly and regularly improved. He had attacks of explosive and involuntary crying when any one came into the room, or when a question was asked, or when he tried to talk much. For the first few days he was often delirious at night, and talked constantly of murder and plots against him, and at times refused medication, saying that he was being poisoned.

At the end of the third week he had quieted down and was free from any distinct irrationality or delusion. He was at that time still unable to talk freely or to speak anything but short sentences. He recognized his family, but did not know where he was. He could not write nor apparently read. He did not remember where he was during the past winter; did not know where he lived, nor where he was at that time; did not know where his children were. He remembered having seen his daughter on that day, but not his wife. He had no appreciation of values, and did not know how much it was costing him to live, or how much could be done with a ten dollar bill. He could sign his name with difficulty. He could not read except isolated words. He could not name objects seen, heard, or felt. He formed correct sentences, but was unable to connect them or make continuous statements. In other words, he was completely disoriented and had amnesia of the past and to some extent of the present. He had a certain form of aphasia, which seemed to be a part of his general amnesia and clouded intelligence. In more technical words, he had apraxia (loss of power to do things because of loss of knowledge how to do them) and agnosia (loss of memory of the use of an object). He did not, for example, know how to use a knife or fork or spoon, and had to be fed, although he had no hemiplegia and he knew the name of the object.

He gradually improved, and in about two weeks was better along nearly all lines. His speech was more coherent, but he soon wandered off and became confused.

On April 29, five weeks after he reached New York, he had got so that he could recall the past fairly well and could tell about his past life. He talked coherently, his explosive sobbing ceased, and his delusions of persecution disappeared.

He gradually regained all his mental functions, so that by the last of May, three months after his illness began, he was able to read and write and converse. All signs of aphasia, agnosia, and apraxia disappeared; his mind was clear, and there was also nothing in his condition which would suggest that he was a sick man, except that his mind worked a little more slowly than usual and was a little more easily fatigued.

In November, 1909, he was seen by me again. He was then well and had no hematoporphyrin in the urine.

Details of Physical Examination. The *hematoporphyrinuria* was first noted about April 1, and it lasted till late in May.

The urine on admission, March 23, showed a specific gravity of 1022; it was of acid reaction, dark amber color; a faint trace of albumin, hyaline and granular casts, and a moderate number of red blood cells were present; indican was increased; there was no sugar; the daily amount was about 1200 c.c. (Dr. Roper).

On April 1, the urine was noted to be a reddish amber color.

On April 9 the reaction to hematoporphyrin was obtained. At this time there were no red blood cells found in the centrifuged specimen. There was still a faint trace of albumin and there were a few hyalogranular and hyaline casts; amount, 1000 c.c.; specific gravity, 1018.

On April 20 all drugs were stopped and he was placed on a meat-free diet for about one week, but the hematoporphyrin continued to be present. It very gradually decreased in amount, yet when the patient was discharged nearly well in May, there was still some present. The trace of albumin became of the faintest, and at the last there were only a few hyaline casts.

At the time when the hematoporphyrin was first found the patient was rather excited, and at times at night delirious, and he thought that people were trying to poison him. This condition cleared up in about a week (April 19).

The blood pressure on admission was nearly 200 mm. Hg. On the second day it was 158, and after this it ranged about 160 to 150; finally, April 20 to 25, it was 140.

The pulse was at first about 90 (84 to 104). After about three weeks it dropped to 72 to 80, and when discharged it was about 70 (66 to 72) (May 3). The temperature was normal.

The respirations were about 20 (16 to 22) and on the first few days were at times of Cheyne-Stokes character.

The Blood Examinations by Dr. Roper. On admission, March 24: Hemoglobin, 90 per cent.; leukocytes, 12,800; polynuclears, 70 per cent.; small lymphocytes, 22 per cent.; large lymphocytes, 5 per cent.; eosinophiles, 2 per cent.; basophiles, 1 per cent. April 12: Hemoglobin, 96 per cent.; leukocytes, 8600; polynuclears, 76 per cent.; small lymphocytes, 19 per cent.; large lymphocytes, 2 per cent.; eosinophiles, 2.5 per cent.; basophiles, 0.5 per cent.

A Wassermann test for lues was made in November, 1909, by Dr. Hastings. It was negative.

Drugs. On admission the patient was given once 15 grains of trional; nitroglycerin, $\frac{1}{150}$ grain every two hours; and potassium iodide, 5 grains three times a day. On March 25 he was given sodium bromide, 15 grains three times a day for one day. On March 26 he was given a 5-grain blue mass pill and Rochelle salt, 1 ounce, and the potassium iodide was increased to 15 grains three times a day. On April 1 he was given 2 compressed cathartic pills. From April 1 to April 20 he received sodium bromide, about 30 grains daily; 2 grains of calomel once; dilute hydrochloric acid, 10 minims three times daily for three days; and a laxative pill—but no other medication, and no sulphonal or other hypnotic. On April 20 all drugs were stopped.

Food. The patient was fed on milk, eggs, bread, stewed fruit, and later, the ordinary hospital dietary of meat and vegetables. He ate normally and moderately. The stools were normal apparently. At one period all meat was excluded from the diet.

Nitrogen Examinations. The total urine under a fixed diet was examined by Dr. Roper five times, and by Drs. Hastings and Warren twice.

The appended table shows the results:

	Total N.	Urea N.	Ammonia N.	Uric or Purin acid N.	Kreatinin N.	Undetermined N.
Hospital Diet,	10.1	8.2	0.4	0.041	0.86	0.6
April 17 to 18,	...	81.2	4.0	0.43	8.5	5.9
1750 c.c.						
April 21 to 22,	8.56	76.0	6.0	2.4	3.4	12.2
1200 c.c.						
Drug free diet,	8.4	6.9	0.3	0.08	0.8	0.32
April 23 to 24,	...	82.15	3.6	0.95	9.5	3.8
1300 c.c.						
Meat-free diet,	6.1	5.1	0.27	0.09	0.49	0.15
April 26 to 27,	...	83.6	4.4	1.5	8.0	2.5
750 c.c.						
Meat-free diet,	6.8	5.7	0.2	0.05	0.61	0.24
April 28 to 29,						
1100 c.c.						
Meat-free diet,	8.1	6.4	0.27	0.13	0.75	0.55
April 29 to 30,	...	79.0	3.3	1.6	9.3	6.8
1320 c.c.						
Meat-free diet,	9.82	83.7	5.2	2.4	5.8	2.9
May 12 to 13,						
1300 c.c.						
Normal	10-14	85-90	3-5	0.9-1.5	3-4	2-6

The upper figures represent the actual amounts, the lower figures the percentage distribution of nitrogen.

Dr. Roper states: "All of these specimens were brownish in color, showed two bands corresponding in position to those of oxyhemoglobin. These were not altered by the addition of glacial acetic acid or reducing agents. No reactions for blood pigment could be obtained. Acidifying with strong HCl changed bands to those of acid hematoporphyrin. Precipitation with BaCl and extraction with acid alcohol after washing with water and absolute alcohol gave a brilliant pink fluid, showing two banded spectrum of acid hematoporphyrin, which changed on making faintly alkaline to the four banded spectrum of alkaline hematoporphyrin. The urine also showed the absorption band of urobilin."

The clinical history would indicate that the patient suffered from an incomplete cerebral thrombosis, involving probably the left frontal lobe. This diagnosis was concurred in by Dr. Allen Starr, who saw the patient in consultation.

The psychosis which resulted was of the confusional and toxic type seen in persons at any age; but the temporary hemiplegia, the agnosia, and apraxia showed that there must have been more than a functional and toxic disturbance.

The study of the blood and urine show that the hematoporphyrin was not the result of a breaking up of blood cells, either living or from a brain clot, for there would then have been some hematin or iron-bearing product. It leads to the conclusion that perhaps the hematoporphyrin in drug cases is due to metabolic (liver?) disorders, and not to a hemolysis, as has been usually supposed.

This view is supported by the fact that in hematoporphyrinuria due to drugs the liver cells show fatty degeneration.

The significance of hematoporphyrinuria apart from drug poisoning and perhaps from intestinal toxemia is not well known.

It has been demonstrated by Garrod and by Soberheim[3] that hematoporphyrinuria may be present in urine after the dark color has disappeared and that, in other words, the color of the urine in this condition is not due to the hematoporphyrin, but to other abnormal pigments. It may, therefore, be that hematoporphyrinuria may be present in the urine without any change in the color. It may be inferred from the present case that its presence is not of so ominous significance as it seems to be in drug cases.

A METABOLIC STUDY OF MYOTONIA ATROPHICA.[1]

WITH THE REPORT OF TWO CASES.

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The case which forms the basis of the following considerations is interesting in several connections: (1) Because it represents a rare clinical entity; (2) because it showed in the light of metabolic study, some findings more marked than is customary in analogous conditions; (3) because one of the findings is now reported for the first time in connection with the disease; and (4) because such metabolic observations seem to throw light on the classification of the condition and suggest a means of arriving at a true diagnosis in obscure diseases of this nature.

A. B., aged thirty-seven years, white, female, married

Chief Complaint. Stiffness in the shoulder, elbow, and knee; pain in the back, and difficulty in walking

Family History. Negative, except that the patient is fairly confident that a sister who died in childbirth had an involvement of the hands similar to hers, but was otherwise healthy.

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[3] Deut. med. Woch., 1892, xviii, 566.

[1] Read at a meeting of the College of Physicians of Philadelphia, December 7, 1910.